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Abstract

Background: The diagnosis of Cerebral Palsy (CP) is often very complicated with earlier diagnoses becoming possible. Primitive Reflexes are automatic, stereotyped movements, directed from the brain stem and executed without cortical involvement. CP can cause primitive reflexes to persist and interfere with the development of normal motor skills. **Case Description:** The patient is a 7-year-old girl who was diagnosed with Dystonic CP at 6 months of age. **Intervention:** An intensive physical therapy bout of 4 days a week for 4 weeks, 3 sessions/week in the clinic for 2 hours each and 1 session/week in the pool for aquatic therapy for 30 minutes for reflex integration and motor function improvement. **Outcome Measures:** The Gross Motor Function Measurement (GMFM) was performed pre and post intervention, along with observation of gait patterns. **Discussion:** This report supports the use of intensive physical therapy which other studies have shown, and the use of reflex integration that requires further research for children with CP.

Keywords: Physical Therapy; Rehabilitation; Primitive reflexes; Reflex integration; Cerebral Palsy; Intensive Physical Therapy

Background

Cerebral palsy (CP) is a permanent disorder of the development of movement and posture that causes activity limitations due to a lesion or abnormality of the developing fetal or infant brain that is non-progressive.[1,2] The diagnosis of CP relies on a combination of neurologic assessment, neuro-imagining findings, and recognition of clinical risk factors. Therefore, diagnosis is often complicated and delayed, and typically occurs at the age of 1 to 2 years or beyond.[1] The diagnosis of CP is made when an infant has evidence of motor dysfunction as well as either abnormal neuro-imagining or a clinical history suggestive of risk.[1] Earlier and accurate diagnosis of CP has become possible in recent years which allows earlier initiation of therapies. Initiating therapies earlier may improve long term outcomes during the period of rapid brain growth and neuroplasticity.[1]

Brain injuries, such as CP, can cause primitive reflexes to persist and interfere with the development of normal motor skills.[3,4] Primitive reflexes are automatic, stereotyped movements, directed from the brain stem and executed without cortical involvement. They develop during gestation, are fully present at birth in term infants, and are elicited readily during the first half-year of life.[3,5,6,7] They are essential for the baby's survival in the first weeks of life, providing important training for later voluntary skills.[6] Each primitive reflex is an involuntary movement with the purpose of stimulating and strengthening a specific sensory-motor neural pathway.[8] During normal development, each primitive reflex emerges sequentially to fulfill a function before being inhibited while the responsibility for continued development is then passed on to the next primitive reflex.[8] Primitive reflexes play a developmental role, preparing the neonate to move against gravity gradually leading to voluntary movement by the process of integration during the first months of life.[3]

Current care for children with CP does not have optimal guidelines regarding frequency, duration, and timing of physical therapy interventions.[9] A common practice is 40-60 minute sessions, 2-3 times a week, although there is no evidence to support the frequency or length of sessions. A large research-practice gap has been reported within the CP field of 30-40% of interventions having no reported evidence-base, and another 20% of interventions provided are ineffectual, unnecessary, or harmful.[10] Broad guidelines have been reported of including interventions that are designed to allow the child with frequent opportunities to practice specific goal tasks. Activities should be developed to allow the child to actively explore their environment and that allow the child to problem solve how to complete tasks. Overall, appropriate interventions have been described as those that focus on exploration, active trial and error hypothesis testing, variability of practice, high frequency of practice, and caregiver education and involvement.[9] While this gives some sort of guideline, it still does not give any specifics on activities to do during sessions. Part of this gray area can be due to every child being different and each family having different goals they want to focus on, which should be the center of treatment plans. Task specific treatments are important and vital successful treatment plans, but often times other confounding variables that may be limiting the child, such as retained primitive reflexes, may go forgotten and therefore limiting the child's progress.

Predictive factors for ambulation in children with CP have been suggested to be divided into three main groups: primitive reflexes and postural reactions, gross motor skills, and the type of CP.[11] A study by Capute et al. showed the lower the number of primitive reflexes present the more likely the child is to be ambulatory and persistent primitive reflexes beyond 12 months of age is an indicator of a poor prognosis.[5,12] It has been shown that retained primitive reflexes affect motor milestones, but more research is needed on how to integrate retained reflexes.[13] The major primitive motor reflexes or patterns that have been described include Moro, palmar and plantar grasp, rooting, sucking, Galant, Asymmetrical Tonic Neck Reflex (ATNR), cross extensor, tonic labyrinthine reflex, and others.[5] This case report will focus on the ATNR, Galant Reflex, and Moro Reflex. The purpose of this case report is to serve as an example of an intensive physical therapy program to integrate primitive reflexes and improvement in Gross Motor Function Movement (GMFM) score.

Case Description

Patient History

The patient is a 7 and a half-year-old female diagnosed with Dystonic Cerebral Palsy. During labor, she was deprived of oxygen for an unknown amount of time causing Hypoxic Ischemic Encephalopathy (HIE) and had 2 seizures following birth. She had low Apgar scores initially; 1 minute: 2, 5 minutes: 3, and 10 minutes: 3. She had an MRI performed showing the basal ganglia, thalamus, and brain stem were involved in HIE. The patient was in the NICU for 5 weeks following birth and was placed on a cooling mat for 5 days to help with brain function. She was diagnosed with CP at 6 months of age and has been receiving Early Intervention therapy at home since birth. The patient was first evaluated in the outpatient pediatric clinic at 7 months of age and has been a patient at the clinic since. At the time of the initial eval patient presented no signs of ATNR, palmar grasp on the left present, symmetrical tonic neck reflex (STNR) present, positive support reflex, plantar reflex present, and able to track objects side to side in sitting and in prone. At 1 year of age ATNR began becoming more prevalent, with being stronger to the left.

The patient is currently receiving speech therapy services at the clinic and occupational therapy at a different clinic in the community. Patient has consistently had therapy services throughout her entire life. The family's goals for this intensive program is to increase patient's independence in ambulation in their new home and to spend more time walking rather than crawling at home.

Diagnosis

The pathways to diagnosis of CP differs depending on whether a child has identifiable risk factors for CP in the neonatal period, which result in earlier screening and closer developmental surveillance.[1] It is recommend that infants with newborn detectable risk factors for CP undergo a standardized neurologic examination, motor assessment, and neuroimaging to help make the diagnosis, ideally before 5 months of age.[1] A review by an international group of CP experts found the best predictive validity for detection of CP before 5 months is neonatal MRI with 86-89% senstivity.[1] Neonatal MRI findings that are particularly suggestive for high risk for CP include white matter injury, injury to deep gray mater structures such as the basal ganglia, cerebellar hemorrhage, and developmental brain malformations. Using MRI at the time of diagnosis of CP can determine the type and extent of brain damage. Risk factors significantly associated with CP in term infants include the following: neonatal respiratory distress syndrome, meconium aspiration, instrumental or emergency cesarean section, birth asphyxia, neonatal seizures, hypoglycemia, and neonatal infections.[1]

Two risk factors this case report will focus on is hypoxic-ischemic encephalopathy (HIE) and neonatal seizures. HIE refers specifically to neonatal encephalopathy resulting from an acute intrapartum hypoxic event. The diagnosis of HIE is made through a combination of neurologic examination findings and biochemical evidence of metabolic acidosis on cord blood gas or arterial blood gas obtained within an hour of birth. Other supporting evidence for HIE may include low Apgar scores, seizures, and brain MRI findings suggestive of an ischemic insult to the watershed areas of the brain or to the deep structures. Recent hypothermia treatment protocols showed a significant improvement in outcomes; however, it is only partially effective, and more research is needed to further reduce the burden of injury. The hypothermia protocols advise systemically cooling patients with HIE within the first 6 hours of life to 34.5° ± 0.5°C for cooling the head, or 33.5° ± 0.5°C for whole body cooling with continued treatment for 48 to 72 hours. Neonatal seizures may occur in infants with HIE but also may be a presenting sign of perinatal stroke of CNS infection.[1]

Four main motor types of CP have been descried in the literature: spastic (by far the most common form, characterized by hypertonicity and muscular resistance to movement), dyskinetic (presents with Athetosis or dystonia), ataxic (primarily presents with difficulties with coordination), and hypotonic (decreased muscle tone). It is important to distinguish between unilateral and bilateral neuromotor involvement because treatment strategies may differ.[1]

Examination

The patient presented to an outpatient pediatric clinic to begin a 4-week intensive physical therapy program ambulating short distances with hand held assist and a reverse walker for long distances. She was classified as Gross Motor Function Classification System (GMFCS) level III because she could walk indoors and outdoors on a flat surface with an assistive device, but when outside on uneven terrain parents carry the patient. The GMFM was performed at the evaluation with the following results: A. Lying & Rolling – 35/51 B. Sitting – 50/60 C. Crawling & Kneeling – 32/42 D. Standing – 11/39 E. Walking, Running, & Jumping – 11/72 and an overall percentage of 54.3%. The patient required use of arms with all tasks in standing or transitioning to standing and is unable to maintain balance when walking without hand held assist. The patient had strong adductor tone and scissored during ambulation.

The GMFM has become the most common functional outcome measure used by rehabilitation specialists to measure gross motor functioning in children with CP over the past 25 years. It is a standardized criterion referenced measurement tool designed to measure gross motor function over time for children ages 5 months to 16 years of age. The GMFM provides outcome scores that reflect how much of an activity a child can accomplish (function) rather than how well the activity is performed. The scores provide an enhanced understanding of activity outcomes ultimately leading towards achievement of contextual participation goals specific to the individual child. The GMFM reliability values range from 0.87 to 0.99.[14] The GMFCS has been widely used internationally to classify level of functional mobility or activity limitations.[15]

Primitive Reflex testing is one of the 4 items of neurological testing recommended in the CP clinical summary.[16] Reflexes tested for this case include: ATNR, Startle reflex, Plantar grasp, and Galant reflex. The presence of ATNR can be tested by placing the child in supine or sitting and turning the child's head to one side. This will cause extension of the extremities on that side and flexion of the contralateral extremities. It is often described as the "fencer" position.[12] The patient's ATNR was retained and was stronger on left than right. The Moro Reflex can elicit the same response from several stimuli; abrupt loud noise near the child's head, sudden movement of supporting surface, or rapidly changing the position of the head from the upright to a backward position. A present reflex results in sudden extension and abduction of upper extremities with opening of the hands as if to grasp, and will freeze in that position momentarily before the arms return to midline.[6,13] The patient's Moro reflex was retained, and stimulated with sudden noise. The Galant Reflex can be tested by scratching the skin of the child's back from the shoulder downwards 2-3 cm lateral to the spinous processes. If the Galant reflex is still present, the child will flex the trunk sideways toward the stimulated side.[5] The patient's Galant reflex was retained and equal on both sides. Plantar grasp is tested by stroking the bottom of the foot up towards the toes and was also demonstrated during ambulation when balance was challenged. The patient's Plantar grasp was retained on the left.

Intervention

The patient was seen for an intensive physical therapy bout of 4 days a week for 4 weeks, 3 sessions/week in the clinic for 2 hours each and 1 session/week in the pool for aquatic therapy for 30 minutes. For all interventions patient was closely guarded since patient's balance was constantly being challenged. For aquatic therapy, patient was in water at hip height and was walking throughout the pool. Patient would continuously reach for therapist to always be touching something in the beginning. Patient then began walking about 5 feet from therapist to student with touching nothing. Distance progressed to all the way across the 30-foot pool. Patient then holding ring with both hands in front of belly button to keep hands down instead of going into high guard position.

In the clinic sessions were based around walking, stairs, and transitions. During sessions, a Togrite strap was donned to facilitate external rotation of her lower extremities to prevent scissoring. During ambulation tasks were targeted to facilitate patient looking ahead and keeping head in midline. Tasks varied from finding certain cards on the wall to reading a sign held in front of her. Patient then began holding items such as two pillow cases filled with items or a large bowl filled with plastic food. This was

to keep her arms at her side during ambulation since she typically brings arms into high guard position. She was then progressed to holding onto the large bowl and walking forward while turning her head to each side. Holding the large bowl prevented her from automatically going into an ATNR position when she rotated her head because it would cause the plastic food to spill out of the bowl if she did, so had to maintain holding the bowl at her belly button. The patient had difficulties with turning while walking so performed infinity walks while holding the large bowl. This also helped the ATNR because she was turning her head to see where she was going when performing infinity walks but had to keep her arms down still. To address the scissoring of gait patient walked through agility ladder that was raised 2 inches above the ground. This facilitated keeping her legs apart and taking big steps since she had to raise her feet high enough to clear the 2-inch boards.

The clinic has a 3-step staircase where the patient can ascend and descend without having to turn around. To begin the stairs were placed where two tensioned cords could be placed horizontal to the ground at the patient's shoulder level. Tensioned cords were used because they give the patient something to hold on to for the feeling of balance but only provide minimal support compared to a handrail since when you lean on the cords they lower to the ground. Patient then progressed to one cord and lastly no cords. Patient was always guarded by two people, one on each side to ensure safety. Patient ascended leading with right and descended leading with left. She could easily bring her right foot up onto the next step, where she had the most difficulties was maintaining single leg balance long enough to push up and bring her left leg onto the step. It often took many attempts before she felt confident in pushing up and had the strength to perform. Patient's toes of left foot significantly curl trying to grip for balance.

Transitions from sit to stand were performed with cues to lean head forward over her toes and to then push up. Patient transitioned to sitting on bench after a repetition of an exercise, required her to turn around and then lower herself to the bench.

The patient's mother was educated on how to stimulate the Galant reflex. While the patient is standing or tall kneeling to run her finger 3 cm to the side from her spine starting level with the top of her shoulder blades all the way down her back to the top of her pants. The mother was instructed to perform this at home as often as possible with the thought that if the reflex keeps getting triggered over and over the patient will become accustomed to it and will no longer trigger the reflex. The family was also instructed on practicing rolling to the right and to the left to help with patient being able to turn when walking. The patient had a lot of difficulties with rolling because she never rolled when she was younger.

Outcomes

At the end of the 4-week plan of care the patient was now ambulating independently. While ambulating, the patient brings hands up into high guard position and with verbal cues can maintain arms at her side. Patient also flexes her neck and tilts her head to the right during ambulation and can bring into midline with verbal cues. GMFM was performed again with the following results: A. Lying & Rolling – 50/51 B. Sitting – 58/60 C. Crawling & Kneeling – 38/42 D. Standing – 26/39 E. Walking, Running, & Jumping – 45/72 and an overall percentage of 82.9%. GMFCS level II because she can walk indoors and outdoors holding on to a railing, but has difficulties walking on uneven terrain and in crowded spaces.

The Minimum Clinically Important Difference (MCID) has been reported based off GMFCS level. Since the patient progressed from level III to level II both MCID's will be reported. MCID's for all dimensions were not found in the literature.[17]

The patient had the following increase in GMFM scores per dimension: A. 15 B. 8 C. 6 D. 15 E. 34. Both dimension D and E had a much larger increase in score than the large MCID for either GMFCS level.

Table 1. MCID of GMFM Dimension D and E for GMFCS II and III.

	GMFCS II		GMFCS III	
	MCID medium (0.5)	MCID large (0.8)	MCID medium (0.5)	MCID large (0.8)
GMFM Dimension D (Standing)	3.3	5.3	1.5	2.4
GMFM Dimension E (Walking, Running, & Jumping)	2.8	4.5	1.8	3.0

*Gross Motor Function Classification System (GMFCS); Minimum Clinically Important Difference (MCID); Gross Motor Measure (GMFM)

Discussion

Other research has shown changes in gross motor function level with most of the children with GMFCS level IV and V at baseline changed to level IV and III respectively, which indicate better improvement in activities in daily life. This study also showed that intensive rehabilitation is beneficial to decrease stiffness and spasm and thereby improve movement in young children with CP, with improvements in self-care and gait. Blocks of intensive therapy can be an alternative to regular dosage of physical therapy but until further studies are conducted, the physical therapy intervention, intensity, and frequency should be tailored to meet the needs of each individual infant and family.[18] This case report is to be used as another support for intensive physical therapy programs.

Primitive reflexes are specifically designed to have a limited lifespan. Once they have completed their developmental functions, these involuntary movements should retire and allow the rational brain to take control over physical movement. However, when a reflex does not fulfill its function fully, it remains active and acts as a signal indicating some neurological weakness.[8] As higher more sophisticated centers of the brain begin to mature, primitive reflexes become a nuisance and must be suppressed for proper neurological organization of the brain to develop during the second six months of life.[7,19] The process consists of the transition from a brain stem reflex response to a cortically controlled response.[3] If the primitive reflexes are retained past the first year of life they can interfere with social, academic, and motor learning.[19] If the primitive survival reflexes remain active beyond 6 months of age they are said to be aberrant, and they represent a structural weakness of the central nervous system, resulting directly from continued brain stem activity at the expense of cortical control.[6]

There are very close links between the inhibition of primary reflexes and the attainment of gross motor milestones in young children.[20] A study by Capute, et al. showed how the presence of primitive reflexes can interact in a significant way with motor activities such as rolling prone to supine, rolling supine to prone, and sitting. Thus, demonstrating the previously assumed association of primitive reflexes and volitional motor activities in normal children. Such associations support the hypothesis that the diminution of primitive reflexes is temporally related to the appearance of volitional motor activity in early infancy.[7]

ATNR has been described as the first use of eye-hand coordination since the arm of the direction the child is looking is extended reaching out in front of the child.[21] This reflex should be integrated by four to six months of age, if retained the child is likely to experience fine and gross motor control problems.[19,20] There may be difficulties with crawling with a fluent reciprocal motion. ATNR affects balance when the child learns to walk since movement of the head to either side will result in straightening of the limbs on one side and upsetting the center of balance.[6] If retained may have difficulties with eye tracking, hand-eye coordination, and with tasks that require crossing the midline.[19]

The Galant Reflex is thought to be important in the birthing process by facilitating movement of the hips as the baby descends into the birthing canal.[19] The Galant Reflex should be integrated by 4 months of age, and if is retained the child may have difficulties siting still, poor posture, and hip rotation on one side when walking.[5,19] If the Galant Reflex is still present once the child is at age of ambulation, can interfere with balance while walking if stimulated.

The Moro reflex, also known as the Startle Reflex, is the baby's most effective alarm system ensuring instantaneous arousal by providing an involuntary and immediate reaction to threat during the period of time the baby is too immature to judge whether that threat is real or not.[6] If the Moro reflex remains present beyond four-six months old the child will remain in a state of heightened awareness and heightened arousal, in a constant fight or flight, where they become easily over-loaded and their ability to categorize information and to form schema is impaired.[5,6,13] The Moro Reflex can affect a child's balance during ambulation due to several stimuli that can cause this reflex, whereas to a person with this reflex integrated would not cause this dramatic of a response.

There is no definitive support for the best approach for Primitive Reflex integration, but a few different approaches to therapy that have been researched include Bimanual training, context-focused therapy, Neurofacilitation of Developmental Reaction (NFDR), and neurodevelopmental therapy (NDT). Bimanual training is repetitive task training with the use of two hands together which improves hand function. Context-focused therapy is changing the task or the environment (but not the child) to promote successful task performance which Improves function. NFDR approach is based on the principles of postural dynamics and coordination dynamics. The intervention has two phases. Phase 1 is composed of preparatory and variability phases. The preparatory component is aimed at normalizing tonal characteristics, promoting mobility, symmetry and alignment, and positioning. The variability component is aimed at eliciting dynamic postural responses, promoting postural stability/task related performances to augment central stability, and reinforce normal motor behavior. Phase 2 is the modulation phase which is aimed at modulation of postural behaviors by altering dynamics and perturbation characteristics. This phase incorporates principles of interaction dynamics for optimizing modulation of dynamic postural behavior using neuro facilitatory contact points, vestibular, proprioceptive, and kinesthetic input recruiting trunk musculature and facilitating postural reactions to give variety of exposure to learn movement experiences temporally and spatially for the development/modification of postural control. NDT is direct, passive handling and guidance to optimize function with conflicting evidence for improved function.[10] NDT is the conventional treatment incorporating positioning, handling at therapeutic key points, using inhibitory and facilitatory techniques such as developmental positions, stretching exercises, etc. and incorporating weight shifting/bearing in developmental positions.[4] This case report used the theory that if you continue to stimulate the primitive reflex, the child will eventually get use to the stimulus and will not have the over exaggerated response of the reflex action.

The purpose of this case report was to present an example of interventions performed in a 4-week intensive physical therapy program to facilitate reflex integration and therefore increase patient's gross motor function. There is a growing body of literature on retained primitive reflexes and gross motor function but are often not tied together in the clinic. Future research in reflex integration in physical therapy is needed to better understand and follow proven activities to integrate retained reflexes. This case serves as a supplement to the limited amount of literature available to physical therapist on reflex integration in intensive physical therapy programs.

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